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Syndrome de Moebius et Syringomyelie : une exceptionnelle association à propos d'un cas et revue de la littérature
Moebius syndrome and syringomyelia An exceptional association Case report and literature review

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Abstract : We report a case of Moebius syndrome associated with syringomyelia in a 6-year-old girl. The association is exceptional. The many physiopathological hypotheses of Moebius have been suggested to explain this syndrome. The hydrosyringomyelic cavity has only been reported in one case. Our case is the second reported in the literature. Several theories have been proposed on the processes involved in the development of the spinal cyst but none is completely compatible with the clinic.

Keywords: Moebius syndrome; Syringomyelia; Spinal cyst

Résumé: Nous rapportons un cas de syndrome de Moebius associé à la syringomyélie chez une fillette de 6 ans. L'association est exceptionnelle. Les nombreuses hypothèses physiopathologiques de Moebius ont été suggérées pour expliquer ce syndrome. La cavité hydrosyringomyélique n'a été signalée que dans un cas. Notre cas est le deuxième rapporté dans la littérature. Plusieurs théories ont été proposées sur les processus impliqués dans le développement du kyste rachidien mais aucune n'est totalement compatible avec la clinique.

Mots-clés: syndrome de Moebius; Syringomyélie; Kyste rachidien.

Moebius syndrome is a rare congenital disorder, the achievement of the seventh and sixth cranial pairs are the rule, the association with the involvement of other cranial pairs (V, X, XI and XII) could cause respiratory disorders. The many physiopathological hypotheses of Moebius have been suggested, including nuclear agenesis or hypoplasia, vascular malformations, and genetic and environmental factors [1].

Case report

This is a girl who is currently six years old. In the sixth month after birth, the parents have noticed the following symptoms: absence of closure of the left eye, strabismus on the same side, feeding problems due to difficulty holding the breast nipple and drooling saliva. In consultation, the physical examination revealed left facial paralysis with a Charles-Bell sign on the left and a left convergent strabismus. There was no other associated oro-facial or musculoskeletal abnormalities. The psychomotor development of the child was normal. The brain scan did not show any abnormality. She was followed by the ophthalmologist for strabismus and reduced visual acuity. The evolution was marked by a regression of strabismus and persistence of facial paralysis.

The recent examination revealed a left Charles-Bell sign, a dysmorphism related to a gap between the jaws with a deviation to the right of the lower mandible (Figure 1). A neurological examination showed a patient without obvious psychomotor disorders.

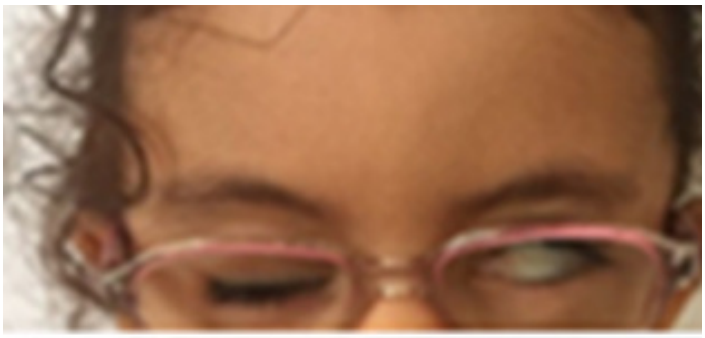

Figure 1.a

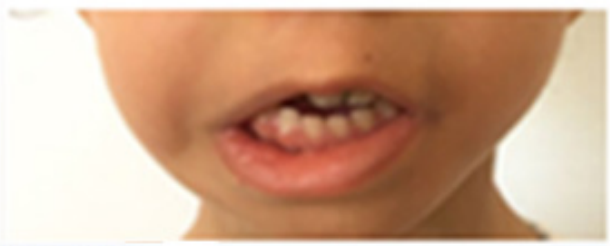

Figure 1.b

Figure 1:
1. a: *Left facial palsy with Charle Bell sign*
1. b: *facial dysmorphism with offset between jaws).*

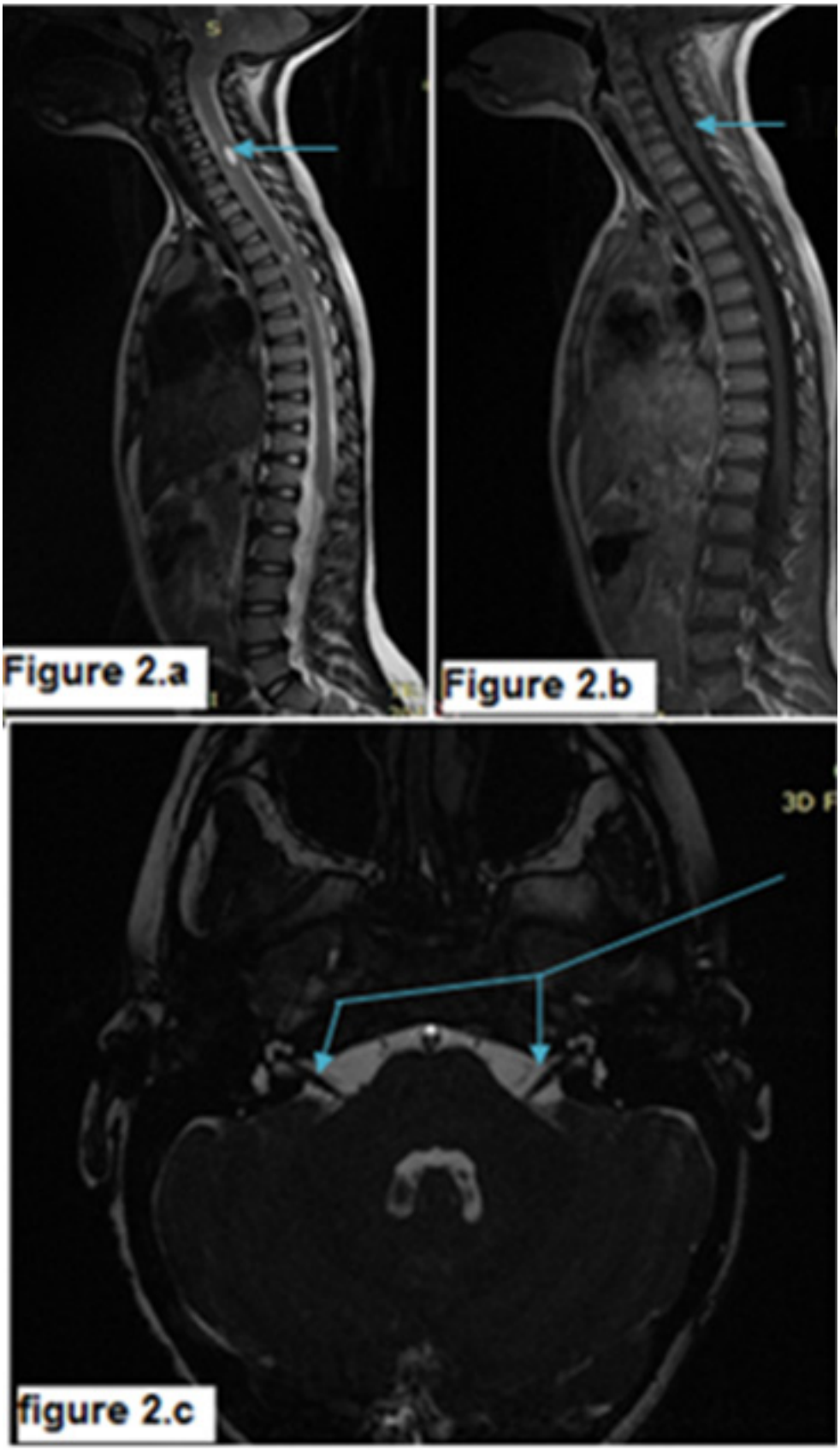

Figure 2 :
2. a: *Cervical syringomyelic cavity in MRI-T2, sagittal section (see arrow).*
2. b: *Cervical syringomyelic cavity in MRI-T1 - Sagittal section.2 (see arrow).*
2. c: *Appearance and normal course of acoustico-facial packs in MRI-axial section (see arrow).*

The fundus retinal examination was normal. The stapedian reflex was abolished on the left side. Cerebro-medullary magnetic resonance imaging (MRI) showed bilateral presence of the cranial nerve VII, with cervical syringomyelic cavity next to C7-T1, there were no other structural abnormalities of the brain (Figure 2). The child had normal hearing and no breathing problems. Radiographic examination of the vertebral column and pelvis revealed no abnormality of the skeleton. The biological assessment was normal. According to these clinical and para-clinical data, the diagnosis of Moebius was retained.

Discussion

The hypothesis widely accepted for its pathogenesis is that of a disruption of the primitive subclavian arteries and their branches before the establishment of a sufficient blood supply on the brainstem by the vertebral arteries. Vascular disruption may be caused by various teratogenic substances acting on the embryo until the end of the first trimester [2]. Genetic mechanisms may also cause hypoplasia or aplasia of cranial nerve nuclei, but they have only a minor role [3].
The hydrosyringomyelic cavity has only been reported in one case [1] . Our patient presented a hydrosyringomyelic cavity that appeared to be asymptomatic. No clear explanation for the development of syringomyelia in this case. In addition to factors directly related to hydrosyringomyelic cavity such as Chiari type I malformation and hydrocephalus, there is no notion of postmeningitic spinal trauma or spinal arachnoiditis in the patient's clinical history.

Several theories have been proposed on the processes involved in the development of the spinal cyst but none is completely compatible with the clinic. Either it is due to pathology of syrinx development [4], or to a cerebrospinal modification of the fluid flow [5]; this has been suggested as an important mechanism for the development of syringomyelia. The physiopathological characteristics of syringomyelia have been extensively studied in the malformations of Bud Chiari (by far the most common association). We believed that displaced brain tissue creates an obstruction to the pulsation of cerebrospinal fluid back and forth through the foramen magnum. The exaggerated pulsatility of the cerebrospinal fluid could lead the cerebrospinal fluid along the perivascular space in the spinal cord; the cavities may develop with relative mechanical weakness [5].

Conclusion

Moebius and syringomyelia patients should be investigated. The association is exceptional. In-depth reports and further research are needed to elucidate this association.

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Declaration of Interests:
The authors declare that they have no interest.
Author contribution:
Hafid Arabi designed the idea and wrote the article. Abdelfettah ELJalil and Ahmed Kharras contributed to the discussion.
Declaration of patient consent:
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed
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Ethical Approval:
No ethical approval was necessary for the writing of this case report. An informed consent form was obtained from the parents of the patient.